

CASE REPORT

PAROTID GLAND VENOLYMPHATIC MALFORMATION PRESENTATION AS MACROGLOSSIA

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Venolymphatic malformations (VLM) are the rare congenital disorders but the parotid gland VLMs are the rarest. Most of the parotid lesions present with unilateral swellings. Aetiology is unknown. Interestingly, this case came in OPD with the macroglossia and only complaint was cosmetic problem. Diagnosis was confirmed on the basis of Magnetic resonance imaging which is gold standard. Doppler ultrasonography showed low flow. Intra lesion electro cautery was done. There is need to focus on malformations and work to find out the causes.

Keywords: Venolymphatic malformation; Parotid gland Venolymphatic malformation; MacroGLOSSIA; Parotid Gland

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INTRODUCTION

Venolymphatic malformations (VLMs) are rare congenital lesions with one case per 2000–4000 live births.¹ Congenital Vascular anomalies were classified by Mulliken and Glowacki in 1982, into; haemangiomas and vascular malformations (VMs). This classification was based on the clinical features and endothelial cell characteristics.² Venolymphatic malformations are differentiated from haemangiomas. Haemangiomas are benign dysplastic lesions which start to grow at birth rapidly. They show regression slowly and spontaneously. On the other hand, VMs are present at birth and continue to grow with the age of patient.³ The International Society for the Study of Vascular Anomalies (ISSVA) classified the vascular malformations on the basis of vessel involved; capillary, venous, arterial and mixed. VLM is a type of mixed vascular malformation. Parotid gland venolymphatic malformations are very rare and 50 cases have been reported in literature before 2014.⁽⁴⁾ This case was

diagnosed with the help of Magnetic Resonance Imaging (MRI) along with Ultrasound and C.T with I.V contrast.

CASE PRESENTATION

A 6-year-old boy presented in outpatient department with the protruded large tongue since birth and visited just for cosmetic reason. He was otherwise healthy, with normal growth and development milestones. He had normal feeding and eating habits since birth. He did not complain for difficulty in breathing, mastication or swallowing. No other significant finding found on systemic enquiry, past medical & surgical history and family history.

On examination, inspection revealed large protruded tongue with ulceration on dorsum and sialorrhea. Angle of mouth was normal. We could not look for crenated tongue due to mouth full of mass. Palpation confirmed soft to firm consistency; approximately 3×5 cm size on measurement outside jaw. No palpable lymph node, cyst, and mass found



Figure-1: Anterior and lateral view

All baselines were within normal limits. Ultrasound revealed large infiltrative cystic lesion from mandibular to submandibular region. On Doppler, no significant vascularity was found. Computed tomography scan (C.T) showed large tongue with multiple cystic areas in superficial and deep cervical spaces which were extending into lower cervical region. While Pre and post intravenous contrast MRI head and neck showed that large infiltrative multilobulated and cystic lesion in submandibular region extending up to lower neck, largest component from left parotid gland. On the basis of MRI, diagnosis of venolymphatic malformation confirmed. Intra lesion submucosal electro-cautery was done in surgery department. It was done using unipolar electrode under local anaesthesia. Tracheostomy and nasogastric intubation were done for optimization of surgery. Patient remained admitted in ward for 15 days and tissue regressed significantly.

DISCUSSION

Parotid gland venolymphatic malformations are low flow benign lesions of head and neck region. Most of the time, these lesions present with unilateral neck swelling by birth and grow with age. Male to female reported ratio is 9:1 and only three cases had extension into parapharyngeal space.³ Head and neck lesions in paediatric population are indication for imaging. Diagnosis can be made antenatal and postnatal as well. Antenatal diagnosis will help obstetricians to decide the mode of delivery and make arrangement for upcoming expected complication. Ultrasound and foetal MR are preferred for antenatal diagnosis. While in postnatal period ultrasound, CT with IV contrast and MRI are suggested to make diagnosis of lesions.⁵ Diagnosis has been easy due to these advanced modalities than before. Cause of disease is unknown but there might be the role of hormone. Definite treatment includes surgical excision but non-surgical options are sclerotherapy, cauterization, radiofrequency ablation and systemic corticosteroids.⁶ Complications due to surgery could be infection, bleeding, scarring and nerve damage which may lead to aphasia for life time.⁷ Systemic use of corticosteroids and

chemotherapy are effective but concerns related to side effects prevail. Few studies suggest the use of Sirolimus for 9–10 months for venolymphatic malformations.⁸

We diagnosed left parotid gland lesion on MRI while clinically macroglossia in this case. He was healthy and active, just presented for cosmetic reason. Intra lesion sub mucosal electrocautery significantly reduced the size of tongue. It has better results with less complication. We did not encounter any complication during procedure or after that.

AUTHORS' CONTRIBUTION

NBK: data collection, data interpretation. KS: Write-up, literature review. FW: Conceptualization of study design. SUK: Literature search, data interpretation. AG: proof reading.

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